MYOPATHY OF THE DISTAL TYPE AND ITS RELATION TO THE NEURAL FORM OF MUSCULAR ATROPHY (CHARCOT-MARIE, TOOTH TYPE).*

By William G. Spiller, M.D.,

PROFESSOR OF NEUROPATHOLOGY AND ASSOCIATE PROFESSOR OF NEUROLOGY IN THE UNIVERSITY OF PENNSYLVANIA.

FROM THE DEPARTMENT OF NEUROLOGY AND THE LABORATORY OF NEUROPATHOLOGY IN THE UNIVERSITY OF PENNSYLVANIA, AND FROM THE PHILADELPHIA GENERAL HOSPITAL.

Various forms of muscular atrophy have been distinguished and sharply separated from one another. While it is possible to make these classifications for typical cases, the borderline cases cause great confusion, and make clinical diagnosis at times extremely difficult.

The primary neurotic muscular atrophy of Charcot-Marie and Tooth was supposed to be so sharply defined that confusion with other types of muscular atrophy could not occur. The characteristics of the type as given by Charcot and Marie[1] are:

Progressive muscular atrophy implicating first the feet and legs, and not appearing in the upper limbs (hands and later forearms) until several years later; the progression of the atrophy being slow.

Relative integrity of the muscles of the limbs near the trunk, or at least, much longer preservation of these than of the muscles of the distal ends of the limbs. Integrity of the muscles of the trunk, shoulders and face.

Fibrillary contractions in the atrophying muscles.

Vasomotor disturbances in the portions of the limbs atrophied.

No pronounced contractions of tendons in the atrophied limbs.

Sensation usually intact, but sometimes affected.

Cramps frequent.

Reaction of degeneration in the atrophying muscles.

Commencement of the affection usually in childhood, the disease often found in several brothers and sisters, and sometimes in the previous generations.

---

*Read before the American Neurological Association, June 4 and 5, 1906.

[1] Charcot and Marie. Revue de médecine, 1886, p. 97.

Charcot and Marie based their conclusions on five cases of their own, and on a few cases in the literature which they believed belong to this type.

I think it well to emphasize the fact that Charcot and Marie in their original paper acknowledged the possibility of implication of the muscles of the limbs near the trunk, at least to some extent. They said that the muscles of the thighs seem to preserve their power and volume during a certain period, but that this integrity often is not real. The vastus internus is first involved. In their summary at the end of their paper they speak of relative integrity of the muscles of the limbs near the trunk, or at least, much longer preservation of these muscles than of those of the distal ends of the limbs. Unquestionably, however, they emphasized the earlier and greater involvement of the muscles at the distal ends of the limbs as the most characteristic feature of the disease.

If we insist on one of the features emphasized by Charcot and Marie; viz., the almost constant absence of all contraction of tendons, many cases classed as progressive neurotic muscular atrophy must be regarded as doubtful.

Cases conforming strictly to the type as described by Charcot and Marie occur, and these probably have a distinct pathology. Such a case is the following, which has been in the Philadelphia General Hospital many years, several times under my care, and so far as I know has not been reported:

N. Hutt, 59 years of age, hostler, was admitted Feb. 15, 1901, at which time the following notes were made:

Chief complaint: Weakness in the arms and legs.

Family history: Father died at about the age of 60 years from some affection of throat, with which he had been ill for one month. Mother died at about the age of 60 years from a second "stroke" of apoplexy. One brother died in infancy and four brothers and three sisters are healthy. The patient is not aware of any family diseases.

He has had only the milder diseases of childhood, and was healthy as a boy and a man, except for the present complaint, and typhoid fever at 17 years. He says he was a moderate user of alcohol, positively denies venereal disease, and has never been married.

History of present illness: He states that at the age of 15 years he began to have pains in the legs and feet, which would last for a week or two, and incapacitate him for work.

These attacks seem to have come on usually in the spring of the year, and he would then be free until the next spring. He had no weakness of the limbs, and was able to work as a hostler, though at about the age of 35 he began to have pain in his hands and arms, and he noticed that he was becoming weak in the legs. These grew thinner and became somewhat deformed. From that time until the present the progress of the disease has been constant, though he has now much less pain than formerly, and only in his feet. During the past five years he has been unable to do any work. The atrophy of the muscles of the

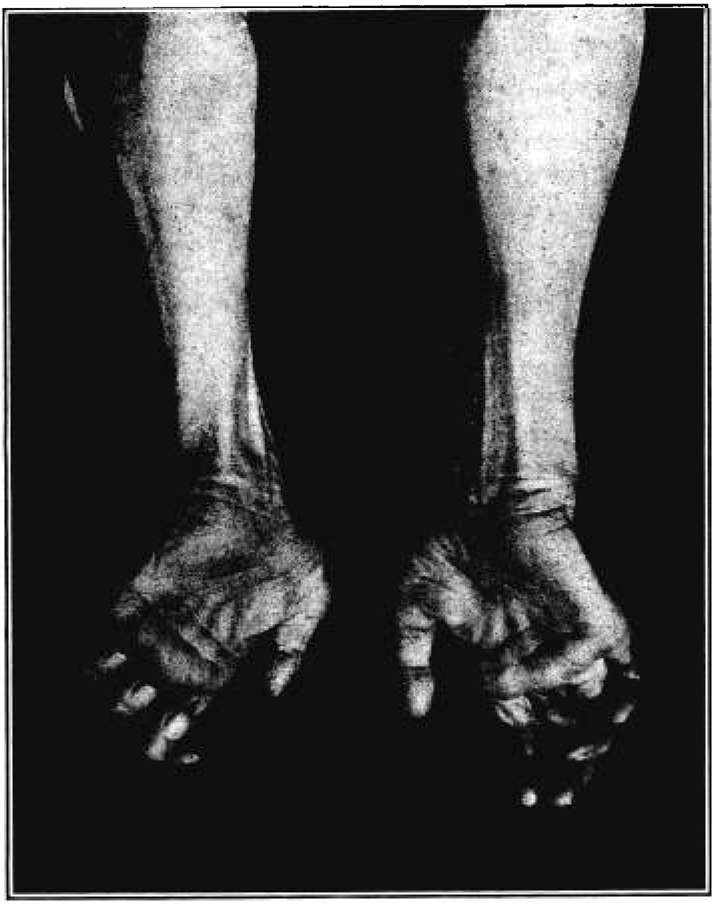

Fig. 1. The neurotic form of muscular atrophy, hands and forearms involved. (Photographed by Dr. Ralph Pemberton.)

legs preceded that of the upper limbs ten years, according to the patient's statement.

He is well nourished, pupils are equal and react to light. Tongue is clean and shows no tremor. Chest and abdomen are well formed, the expansion of the chest is fair, lungs are normal, heart sounds are muffled and distant.

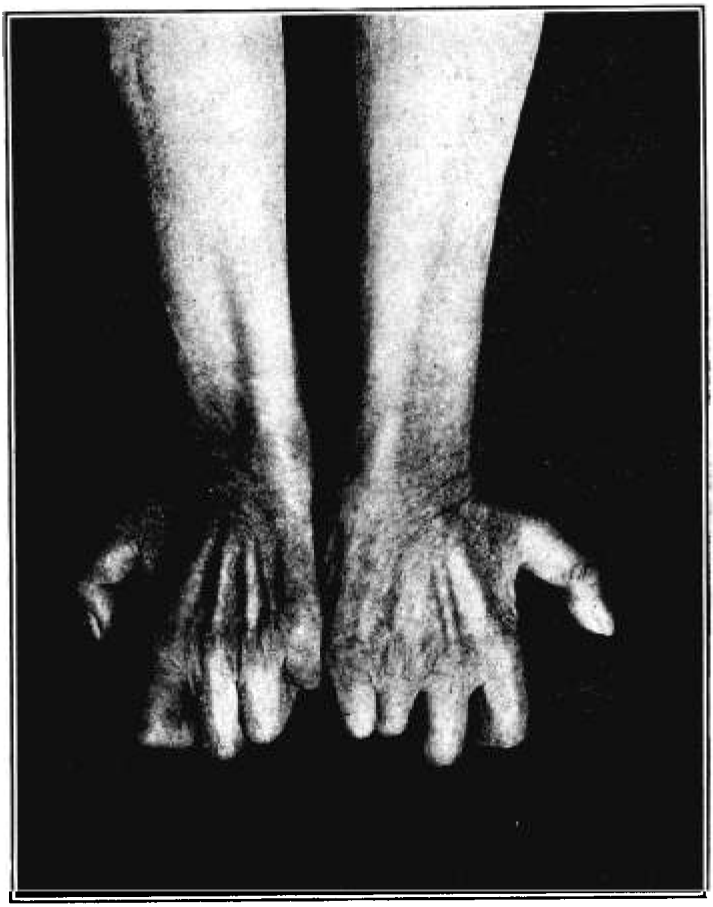

Fig. 2. The neurotic form of muscular atrophy, hands and forearms involved. (Photographed by Dr. Ralph Pemberton.)

Examination of eyes by Dr. Shumway, Oct. 20, 1904: The pupils are equal, respond promptly; eye movements are good. Ophthalmoscopic examination: Haziness of lens, retinal veins full and show pressure signs, result of arteriosclerosis, otherwise fundus is normal. Vision, O. D. 5-8; O. S., 5-10; fields are normal.

Condition of the patient at my examination March 7, 1906: The atrophy is intense in the hands (figs. 1 and 2), distinct in the forearms, but the arms above the elbows are well developed. Sensations for touch, pain and temperature are normal in the upper limbs. Sense of position is normal in the fingers. The grasp of each hand is very feeble. The muscles of the trunk and head are well developed. He raises the upper limbs above the head with much power, and is able to dress himself and handle his clothing with considerable skill, notwithstanding

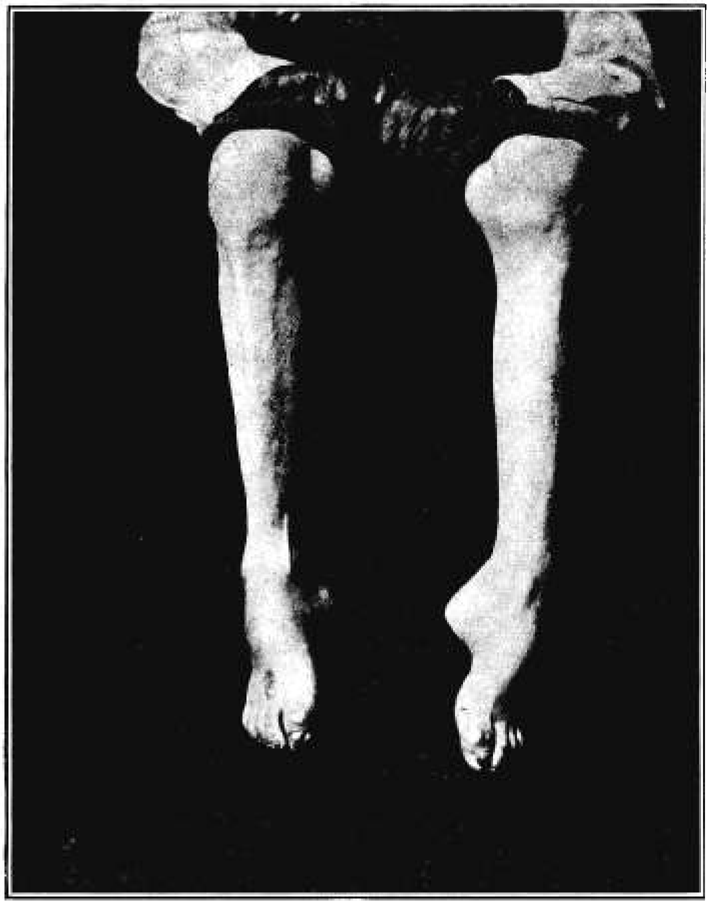

Fig. 3. The neurotic form of muscular atrophy, feet and legs (not thighs) involved. (Photographed by Dr. Ralph Pemberton.)

the intense atrophy of the hands. Biceps and triceps reflexes are weak.

The lower limbs, above the knees, are well developed, and if there is any atrophy here it is very slight. Below the knees the atrophy is marked. The feet are inverted and talipes equinovarus (fig. 3) is very pronounced on each side. The patellar reflex and Achilles tendon reflex are lost on each side. The toes are somewhat cyanotic. Sensations for pain, touch and temperature are normal in the lower limbs. He is able to walk without assistance and without crutch or cane, but his gait is rather slow, with legs far apart and his feet turned inward. His gait is somewhat steppage in character.

If we hold to the type of muscular atrophy as presented by this patient; viz., atrophy confined strictly to the distal parts of the limbs, we shall be able to recognize the neurotic muscular atrophy as a distinct type, with a distinct pathology. The cases of this kind with necropsy are very rare, we are obliged to depend on the findings of Marinesco, Sainton, and Dejerine and Armand-Delille. Oppenheim includes the cases of Dubreuilh and Siemerling, but Sainton regards the former as far from typical, and does not refer to the latter, which should, I think, be excluded.

In Siemerling's[2] case there was great atrophy of all the extremities in the upper parts as well as in the lower, and of the trunk; flaccid complete paralysis of the lower limbs, so that all voluntary movements of these limbs were lost. Siemerling found degeneration of the posterior and lateral columns, most intense in the lower thoracic and upper lumbar regions, degeneration of peripheral nerves and muscles; atrophy of the cells of the anterior horns, of the columns of Clarke, of the anterior roots, and of the spinal ganglia.

He thinks there can be no doubt that the case belongs to the spinal neurotic atrophy, but if we accept this opinion we cannot limit the type to those cases in which the atrophy affects the distal portions of the limbs, and the proximal portions and trunk escape.

In the case of the neurotic muscular atrophy studied by Paul Sainton[3] the lesions were:

Sclerosis of the posterior columns, especially of the columns of Burdach. Slight degeneration of both pyramidal tracts. Alteration of the columns of Clarke. Atrophy of the cells of the anterior horns. Slight degeneration of the intramuscular nerves. Slight sclerosis of the nerves of the forearms and legs, very distinct only in the peroneal nerves. Atrophy of the muscle fibers even causing complete disappearance of some fibers with proliferation of connective tissue.

---

[2] Siemerling Archiv. für Psychiatrie, Vol. 31, 1898, p. 105.

[3] Paul Sainton Nouvelle Iconographie de la Salpêtrière, Vol. 12, 1899.

In this case the atrophy began in the upper limbs. The lesions resembled those observed by Marinesco in his case, except that Marinesco found the anterolateral columns intact. Judging from these two cases Sainton regards as the lesions of this form of muscular atrophy: sclerosis of the columns of Burdach and Goll, atrophy of the cells of the anterior horns, and alterations of the peripheral nerves of varying intensity, sometimes slight.

.The lesions found by Dejerine and Armand-Delille[4] in a case of neurotic muscular atrophy were degeneration of the posterior columns, degeneration of some of the nerve cells of the anterior horns of the cervical and lumbar regions without diminution in the number, chronic meningitis, degeneration of the muscles of the hands and feet, alteration of the intramuscular nerve fibers in the hands and feet (*i.e.*, many nerve fibers of small size, many empty nerve sheaths and a few nerve fibers in the process of degeneration). The nerve trunks, the cutaneous sensory nerves, and the anterior and posterior nerve roots with slight exception were normal.

The literature contains many cases reported as belonging to neurotic muscular atrophy in which some atypical features were present. Sachs'[5] two cases were probably the first reported in America. In his first case the patient had pronounced kyphosis of the lumbo-sacral region, slight wasting of the infraspinatus muscle, and decided diminution in the power of thighs as well as of legs.

The younger brother of the first patient had general emaciation of all parts of the upper extremities, very distinct atrophy of the infraspinatus and marked general atrophy of the legs, which seems to imply involvement of the thighs.

In discussing the cases together Sachs says the atrophy caused a weakness of the thigh muscles as well as of the muscles of the legs, and he speaks of atrophy involving all the muscles of the lower extremities as common in the neurotic muscular atrophy.

---

[4] Dejerine and Armand-Delille. Revue Neurologique, 1903, p. 1198.
[5] Sachs, Brain, 1890, Vol. 12, p. 447.

In one of Hoffmann's[6] cases the lower limbs were wasted in all parts, and chiefly below the knees, but the strength of the thigh muscles was good. Fibrillary tremors were not observed. In another case, Hoffmann found wasting of the upper part of the left trapezius muscle and flattening of the supraspinous and infraspinous fossæ, without any loss of function. Fibrillary tremors were not seen. In another case he says the thigh muscles were distinctly weak and much wasted, especially in the lower part of the thigh. The thoracic muscles were poorly developed and the ribs were prominent. In still another case all the thigh muscles were much atrophied, as well as the muscles of the buttocks and the lower part of the extensors of the back. The patient had a distinct lumbosacral kyphosis, and experienced difficulty in rising from the ground, such as is seen in progressive muscular dystrophy. The legs were kept far apart and the patient climbed upon himself as in muscular dystrophy.

Hoffmann accepts involvement of the facial muscles as a part of the neurotic atrophy.

In the case reported by George W. Jacoby,[7] and recorded as one of progressive muscular atrophy of the peroneal type, severe pain was felt in the lower extremities when the child was two years old. When she was four years old one leg was found to be weaker and thinner than the other. Later atrophy of the thigh on the side opposite to that of the affected leg was noticed. There was marked lordosis and slight lateral curvature. The muscles of both thighs were in a continual state of unrest, almost like fibrillary twitchings. The right foot was in equinovarus position. There were no sensory disturbances, but there was partial reaction of degeneration in the affected muscles. The arms were not affected.

Jacoby acknowledged that the case was not typical, inasmuch as the atrophy, although bilateral, was asymmetrical, one leg being affected and the opposite thigh.

---

[6] Hoffmann. Deutsche Zeitschrift für Nervenheilkunde, Vol. 1, p. 95.
[7] G. W. Jacoby, JOURNAL OF NERVOUS AND MENTAL DISEASE, 1894, p. 259.

Sachs, however, concurred in the diagnosis of muscular atrophy of the peroneal type.

In at least one of Diller's[8] two cases (brother and sister), the thighs also were wasted, and the patient climbed upon his lower limbs as is seen in muscular dystrophy. He had left talipes equinus. Response to electricity in the paralyzed legs was very sluggish and reaction of degeneration was obtained.

In the first of the two cases reported by Given Campbell[9] the disease progressed in the lower limbs until these limbs became useless and the muscles of the thighs also were much wasted. The deltoid muscles, especially the left were weak. The weakness and wasting developed hand in hand, and began in the peroneal group of muscles. The small muscles of the hands were not affected. The muscles of the trunk were not wasted, unless possibly the abdominal muscles to a slight extent.

I could add to these cases others to show that atypical features have not prevented the diagnosis of muscular atrophy of the Charcot-Marie-Tooth type, and therefore it is not surprising that cases of muscular dystrophy have been diagnosed clinically as belonging to the type. In this connection it is interesting to read Sachs' words published in 1889 (Brain) when the peroneal type of muscular atrophy was little known. He said: "There is a consensus of opinion, however, on this one point, that the cases in question do not belong to the category of primary muscular dystrophies." He was speaking of the peroneal type of atrophy. Oppenheim and Cassirer[10] showed that this consensus of opinion no longer exists.

They pointed out that the clinical picture of the neurotic muscular atrophy is not so sharp as Hoffmann believed, and that the pathology is still uncertain. In their case the muscular atrophy had existed about two years. The symptoms began with severe pain in the lower and upper limbs. The distal portions of the lower limbs, especially those innervated by the peroneal nerves, were affected, and in the upper limbs only a part of the small hand muscles, and later the triceps and supinator longus, were implicated. In some muscles incomplete reaction of degeneration was obtained, in others all electrical response failed. The tendon reflexes were diminished. The orbicularis palpebrarum on each side was involved. Later the calf muscles became atrophied. The diagnosis seemed to lie between chronic multiple neuritis and neurotic muscular atrophy. The central and peripheral nervous systems were normal, but the muscles were much degenerated.

---

[8] Diller. Philadelphia Medical Journal, March 17, 1900, p. 642.

[9] Campbell. JOURNAL OF NERVOUS AND MENTAL DISEASE, 1900, p. 274.

[10] Oppenheim and Cassirer. Deutsche Zeitschrift für Nervenheilkunde. Vol. 11, p. 143.

As regards the involvement of the orbicularis palpebrarum, Oppenheim and Cassirer refer to the fact that in Sachs' cases the infraspinati were atrophied, in a case of Hoffmann's the upper portion of the left trapezius and the supraspinati and infraspinati were atrophied, in a case of Donkin's reaction of degeneration was obtained in the left trapezius, in a case of Hülemann's the pectoral, rhomboid, latissimus dorsi muscles were somewhat atrophied, in a case of Eisenlohr's the atrophy of the upper limbs was like that of myopathy, but of the lower limbs like that of the neurotic musclar atrophy. All these cases were regarded as belonging to neurotic muscular atrophy, and yet they showed some features in common with muscular dystrophy. Especially interesting are two cases in sisters observed by Dähnhardt, in one, the wasting resembled the neurotic muscular atrophy, in the other the progressive muscular dystrophy.

Oppenheim and Cassirer concluded that the clinical picture of progressive neurotic muscular atrophy may be caused by muscular dystrophy, and that the pathology of the former is not always the same and not as yet clearly defined.

Gowers[11], in reporting a clinical case of distal myopathy in 1902, said he had not seen a similar case nor did he know that one had been recorded. The patient, a boy, was eighteen years old, and was the only member of the family affected. The symptoms first attracted notice when he was ten or twelve years old; then it was noticed that he often caught his toes against the ground in walking. At a later date his hands were found to be weak. The feebleness of hands and feet steadily increased until it became pronounced. He was unable to flex the ankles, although he could just extend the toes and could move each foot slightly in and out by the tibialis anticus and the peronei. He could extend the ankle joint by the calf muscles with some force. The movements of the knee and hip were performed with good power, the knee jerks were present, but the left was less than the right. The thigh muscles were of normal size; the anterior tibial muscles were distinctly smaller on the left side; the calves were rather large and firm, and were sufficiently large to suggest a resemblance to pseudo-hypertrophic paralysis. The grasp was extremely feeble, with the right hand he could not move the dynamometer, and with the left he could only move the index to 2 k° instead of 50 or 60. Extension was also feeble; he could get the fingers with the wrist into line with the forearm, but could not fully extend them when the wrist was overextended. The muscles of the forearms and hands were small, but presented no wasting comparable to that which is seen in progressive spinal atrophy. Above the forearm the muscles had fair power and presented no wasting; only a trifling atrophy of the middle part of the trapezii could be observed. Electrical excitability was lowered in the affected muscles in proportion to their feebleness, and equally to faradism and voltaism.

---

[11] Gowers. British Medical Journal, July 12, 1902.

The neck muscles were normal except the sterno-mastoids. He could not raise the eyebrows at all, and closure of the eyes by the orbicularis muscles were weaker than normal.

Gowers said this case differed from all recognized forms in the purely distal distribution of the affection in the limbs and the normal state of the muscles near the trunk. The case resembled therefore the neurotic muscular atrophy, but the implication of the sterno-mastoid, frontalis and orbicularis palpebrarum muscles probably prevented Gowers from classing the case under this head.

In the case of myopathy of the Aran Duchenne type reported by Dejerine and Thomas,[12] the muscular atrophy was confined to the upper limbs and the hands were much wasted. The muscles of the head, neck, face, trunk and lower limbs were not at all affected. Fibrillary contractions were seen in the upper limbs. The symptoms began when the patient was 49 years old, and lasted thirty-one years. It is not surprising therefore that the case was regarded as one of myelopathy during the lifetime of the patient. The spinal cord and nerves, even the intramuscular nerves, were normal. The muscles were much altered, many muscular fibers had disappeared, and those that remained were much atrophied, many had lost the transverse and longitudinal striations, the sarcolemma nuclei were multiplied and some were enlarged.

---

[12] Dejerine and Thomas. Revue Neurologique, No. 24, Dec. 30, 1904.

The case of muscular dystrophy affecting the hands and feet reported by C. Macfie Campbell[13] was without necropsy. Other members of the family were similarly affected. Fibrillary twitching in the interossei and short flexor of the thumb, and reaction of degeneration in two of the atrophied muscles of the lower extremities raises the suspicion that the case possibly may belong to the type of neurotic muscular atrophy.

These are the only instances in literature I know of where muscular dystrophy gave distinctly the clinical signs of neurotic muscular atrophy, but a case of this character with necropsy has come under my observation. I hesitated during the lifetime of the patient to make the diagnois of neurotic atrophy, although the case was regarded by some as belonging to this type. I hesitated because the thighs and upper arms and trunk were also much atrophied, and I felt that the diagnosis should be left open. The commencement of the atrophy in the lower part of the lower limbs, the slight reaction of degeneration in these parts, the talipes equino-varus on each side, the symmetry and the later involvement of the upper limbs suggested the neurotic form. The microscopical examination has shown a normal central and peripheral nervous system with much atrophy and disappearance of muscle fibers.

G. R., age 28 years, was admitted to the Philadelphia General Hospital Oct. 11, 1898, complaining of pain about the knee and difficulty in walking.

Family history: Father died of heart disease at about 25 to 30 years of age, mother living and well, at 50 years of age.

The patient was married at 23 years or age. He has one child, three years old, in good health. He uses neither tobacco nor alcoholic drinks, and has been employed as a watch case engraver since his fifteenth year.

Past medical history: He has had childhood diseases. When ten years old he fell and injured his back and could not walk for two months (the condition was diagnosed as spinal meningitis), but he recovered the use of his limbs satisfactorily in about one month after getting up. Three years ago he had an attack of cystitis.

History of present illness: About two years ago he fell from a street car, striking on his hip and back of his head, badly bruising his right hip and thigh. He suffered no particular inconvenience from this at the time, except a slight but constant pain in the knee joint. About six months later, upon arising one morning, he fell to the floor and found himself unable to walk, but this was probably not the first manifestation of weakness. Two or three days later he was able to walk with a cane, and after a few days could walk without it. He walked without a cane about two weeks. During this time, while walking along the street, he fell and had to be assisted to a car, and could not walk during the remainder of the day. About a week after this he was obliged to use the cane again, and has continued doing so since. For the past year and a half the inability to walk has steadily progressed, and coincident with this has been the muscular wasting. No involvement of bladder has occurred, except a cystitis three years ago, previous to his accident. He had noticed weakness first in the left foot, later in the right foot, and three months still later his upper limbs became affected. He stated that the wasting began in the feet, extended to the legs and thighs, and that the upper limbs were affected first in the distal parts.

---

[13] Campbell. Review of Neurology and Psychiatry, March, 1906, p. 192.

Condition upon admission: He is a somewhat poorly nourished adult male, of above average height; pupils are dilated, the left more so than the right, both react to light, accommodation and convergence. Speech is normal. Tongue protrudes straight and shows no tremor. Pulse is small, quite irregular; arteries are soft.

Chest and lungs: Chest is somewhat emaciated; expansion is good. Lungs are normal.

Heart: Apex beat is visible and palpable in the 6th interspace, 1½ inches within the nipple line. First sound is loud and strong; second sounds are both accentuated. Liver and spleen are normal.

Arms: Grip, right hand, is 21; left is 17. The arms are emaciated, but not atrophied. Ataxia is not detected.

Legs: Marked general wasting of the muscles, apparently equal in both legs. Foot-drop is marked. Toes are slightly flexed. Spasticity of legs is not present.

Reflexes: Knee jerks are absent; no ankle or patellar clonus; superficial reflexes are present. Tactile sensation is preserved and normal. Pain and temperature sensations are normal.

Nov. 7, 1898: Patient's physical condition is improved since admission. No change in walking, or in rising from chair. He is gaining slightly in weight, but complains of some pain in the legs and ankles.

An examination of the eyes April 14, 1899, by Dr. Charles A. Oliver showed healthy eyegrounds, paresis of left external rectus and left superior oblique, with diplopia, which had been present a few days. This was not persistent.

At this time he was unable to walk, and could not stand alone, although he could move his legs somewhat. Foot drop was marked on each side. The legs could not be extended. Muscular atrophy had been progressive.

On June 25, 1899, a note was made that the patient could walk, but had inversion of the foot on attempting to walk.

Sept. 10, 1899, the patient was granted a two-days pass on which he went out Friday. He was brought back by a patrol wagon last night, having been picked up at Market St. ferry, where he had suddenly lost all power of his legs. When admitted he was complaining of abdominal and lumbar pain, loss of power in legs as well as a feeling of complete exhaustion. This evening he is much better, and already can move his legs fairly well, has lost much of the lumbar and abdominal pain, and feeling of exhaustion.

Sept. 26, 1899. He has improved rapidly since his return; he can walk, but is still kept in bed.

April 13, 1901. Examination by Dr. Pearce. Both upper extremities, both above and below the elbow, respond quickly and well to faradism. There is no reaction of degeneration. Elbow jerks and biceps jerks are present, but much diminished. There is no localized wasting in upper extremities. In the lower extremities there is marked quantitative change below the knees, and very slight qualitative change in the anterior and posterior groups of muscles. There is no sensory change anywhere. Knee jerks are absent. Plantar reflex is present. No arthropathies; pupillary reactions are normal.

July 19, 1903. He has not walked for two and a half years. Sensations of touch and pain are normal.

Aug. 20, 1903. Dictated by Dr. Weisenburg. Patient is unable to walk; he can flex the right thigh on the hip slightly; left thigh movement is very poor. No movements possible in ankles; he can flex or extend the toes of both feet very slightly. Both feet are in a position of talipes equino-varus, the left more than the right. Both feet are cold. Movements of both upper limbs are free in all directions. Grip in the hands is almost nil. No power in elbows at all. Power in shoulders fairly good. Both upper extremities, shoulders, back and chest are wasted; abdomen not so much. Wasting is about equal on both sides. Thenar and hypothenar eminences on both hands are almost absent. Atrophy of interossei is marked. In the forearm the flexor group is more atrophied than the extensor; the biceps is atrophied on each side. Deltoid is slightly atrophied. Pectoral muscles wasted, also the interossei on both sides. Supraclavicular and infraclavicular fossae well marked, also interscapular region. All muscles of the shoulder girdle are atrophied, including the latissimus dorsi on each side. No fibrillary twitching is noticed in the muscles. All reflexes in the upper limbs are lost.

He has never had any urinary or rectal disturbances. The pupils are equal, respond to light, accommodation and convergence. No apparent weakness of left internal rectus or left superior oblique. No involvement of 5th or 7th nerves. Tongue protruded straight; no tremor or atrophy. No atrophy of the muscles of the face or neck.

At present the wasting in both lower extremities is about equal. The femoral group on each side is very much wasted; the calf muscles are wasted, as are the thigh muscles of each side. All the reflexes in the lower extremities are lost. Plantar irritation produces flexion of all the toes. Abdominal and cremasteric reflexes are present and active.

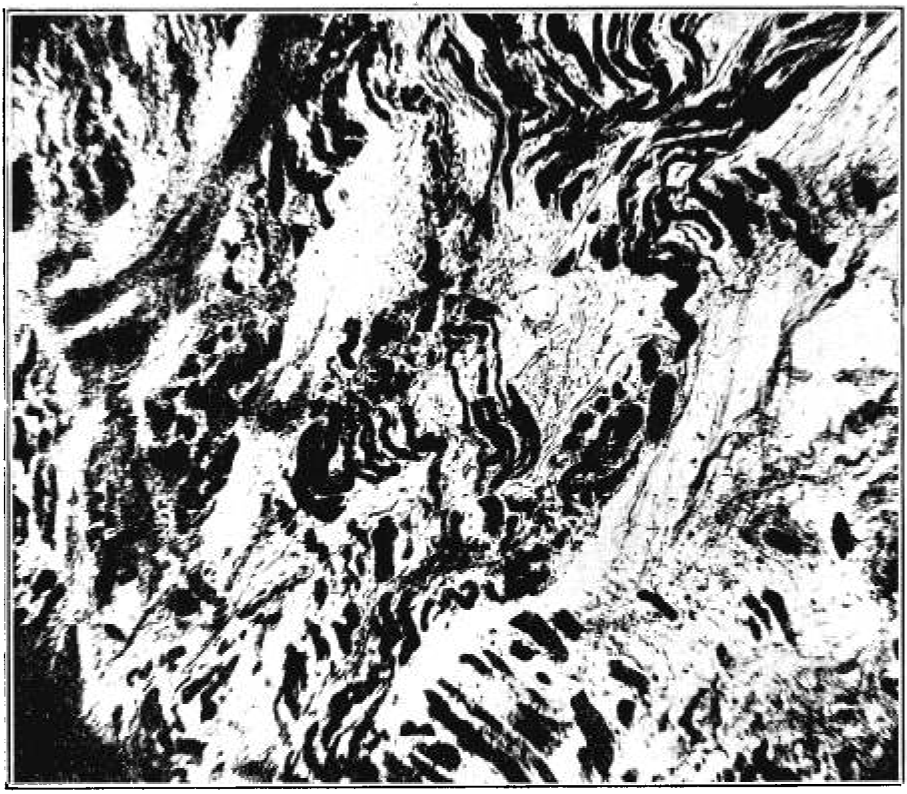

Fig. 4. Piece of muscle from the sole of the foot in a case of myopathy of the distal type. (Photographed by Dr. Alfred Reginald Allen.)

He says he has dull pains in the legs and arms, but has never had any sharp shooting pains, nor any sensory disturbances like pins and needles. Sensation is apparently normal.

Oct. 12, 1904. Dr. Shumway: Right vision, 5-5; left vision, 5-6. External ocular movements are normal. Pupil reaction is prompt. No diplopia in any part of the field. Fields full; perfectly normal eyegrounds.

July 21, 1905. Examination by Dr. Pickett: Ankles are almost fixed in position of foot drop. The right knee cannot be extended completely. Strongest movements of the body are extension of the feet (plantar flexion), supination of the forearm, then after this pronation and extension of wrist and fingers. Preserved supinators and extensors make the forearm bulge as if it and the bones were bent. His movements in dressing are at times so irregular by the unequal strength of the different muscles as to constitute a kind of ataxia.

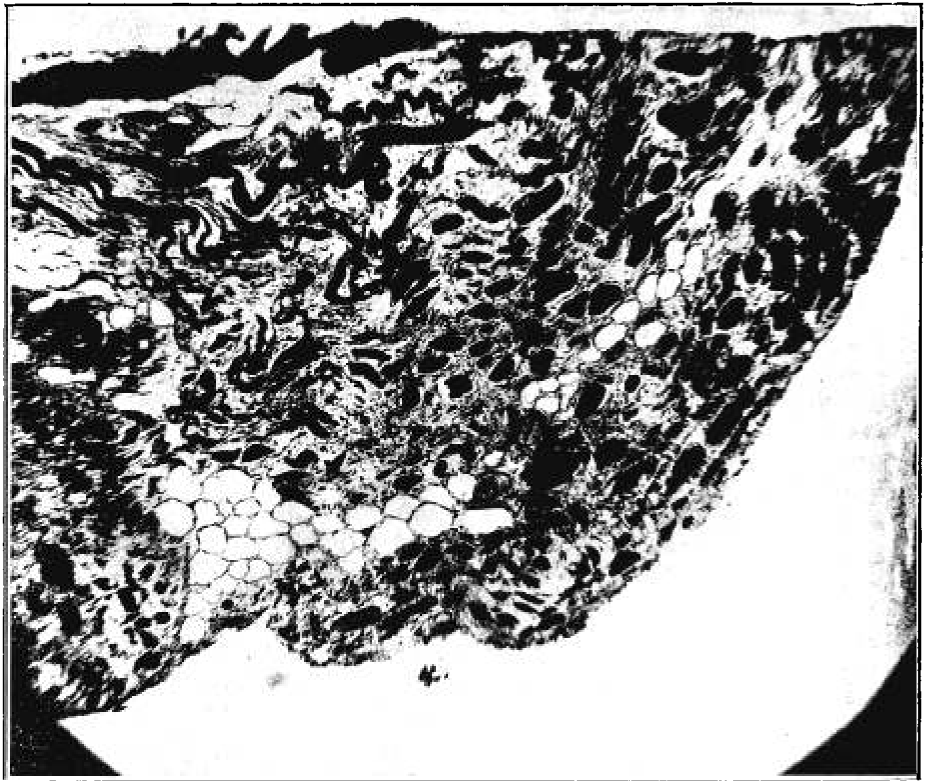

Fig. 5. A piece of muscle from the sole of the foot in a case of myopathy of the distal type. (Photographed by Dr. Alfred Reginald Allen.)

Dec. 6, 1905. Patient's nervous condition is the same as described above. He has a cough and expectorates moderately;

has night sweats, and the sputum shows the presence of tubercle bacilli.

Urine analysis: Dec. 6, 1905. Yellowish; slight sediment; acid reaction; Sp. Gr. 1010. No sugar or albumin. Microscopic: red blood cells, a few; few calcium oxylates, epithelial cells and debris.

He died Jan. 11, 1906.

This man was examined frequently and lectured upon by me several times. He was repeatedly in my charge at the hospital. His condition at the time of death was as follows:

The lower and upper limbs were greatly atrophied and equally so at all parts. Contracture at the right knee was present, so that the leg could not be fully extended upon the thigh. The left leg could be fully extended on the thigh. There seemed to be contracture at the right hip. Talipes equino-varus was present on each side. No contractures were observed in the upper limbs. Emaciation was shown also in the muscles of the trunk and face. Slight lordosis was present in the lumbo-sacral region, and there was some protrusion of abdomen, probably secondary to the lordosis. There was no evidence at the necropsy of old spinal injury, and excepting the lordosis, there was no deformity of the vertebræ. The wasting of the face may have been caused by tuberculosis, as it was not present in 1903.

Sections from the cervical and lumbar regions of the spinal cord stained by the hematoxylin method of Weigert or the method of Marchi, by acid fuchsine, hemalum or thionin are normal. A piece of muscle from the foot (fig. 4 and 5) shows no recent degeneration by the Marchi method, but the long-standing degeneration is pronounced, and is better shown by other methods of staining. The fibrous and fatty connective tissues are much increased, the muscle fibers are greatly atrophied. The longitudinal and transverse striations are well preserved, and the sarcolemma nuclei are increased in number in some of the muscle fibers. Nerve bundles between the muscle fibers stain well by the Weigert hematoxylin stain, as do also sections of one of the plantar nerves. No degeneration is seen by the Marchi method in the latter, but the blood vessels of the nerve are thickened. A piece of muscle from the back of the trunk appears normal.